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Herniation to foramen magnum in the course of cerebellitis in a 4-year-old boy, as shown by CT and MRI – case report

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Summary

Background:

Acute cerebellitis is an uncommon but dangerous complication of infectious diseases. Besides neurological examination, neuroimaging (especially MR imaging) is very useful for diagnosing cerebellitis.

Case Report:

A 4-year-old boy was admitted to the hospital with a 1-week history of fever, vomiting and headache. His past medical history was unremarkable. The physical examination revealed disturbance of consciousness and truncal ataxia. He underwent urgent CT and MRI examinations which demonstrated isolated swelling of the cerebellar hemispheres and the vermis, with increased signal intensity in T2-weighted, FLAIR, and DWI sequence and a significant mass effect associated with tonsillar herniation. An emergent life-saving suboccipital craniectomy was performed with removal of the C1 vertebral arch. There was a gradual clinical improvement, and a follow-up brain MRI revealed disappearance of cerebellar swelling and of mass effect.

Conclusions:

Magnetic resonance (MR), including diffusion-weighted imaging (DWI) sequence, plays an important role in the diagnostic work-up of cerebellitis in children. This imaging method is very useful for detecting cerebellitis, evaluating its severity and monitoring the disease.

Key words:

cerebellitis • herniation • MRI

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Background

Acute cerebellitis is a rare syndrome characterised by a sudden onset of symptoms of the cerebellar damage. The disease may follow from a primary or secondary infection, or may be a result of postvaccinal reaction. It concerns mostly children at the age of 6 [1]. Cerebellitis is usually a benign, self-limited disease [2]. However, it may have a fulminant course and lead to death [3] due to upward or downward herniation. It may also cause a severe atrophy of the cerebellum [4]. Owing to that, early diagnosis is essential for implementing appropriate treatment.

Case Report

A 4.5-year-old boy without any remarkable medical history was admitted as an emergency case to a District Hospital due to a 1-week history of vomiting, fever, headache, disturbance of consciousness in the form of pathological sleepiness, and disturbed coordination of movement (especially the gait). A general examination of the cerebrospinal fluid showed an elevated white cell count of 11 cell/mm³ (normal range: 0–5 cell/mm³). No bacterial antigens were detected. A contrast-enhanced CT revealed normal brain and intracranial fluid spaces. Ophthalmoscopy did not reveal any changes in the fundus of the eyes. Viral aetiology was assumed. Treatment with Aciclovir was started.



Figure 1. Contrast-enhanced brain CT showing a diffuse cerebellar swelling with a significant mass effect – compression of the fourth ventricle and obliteration of the quadrigeminal cistern.

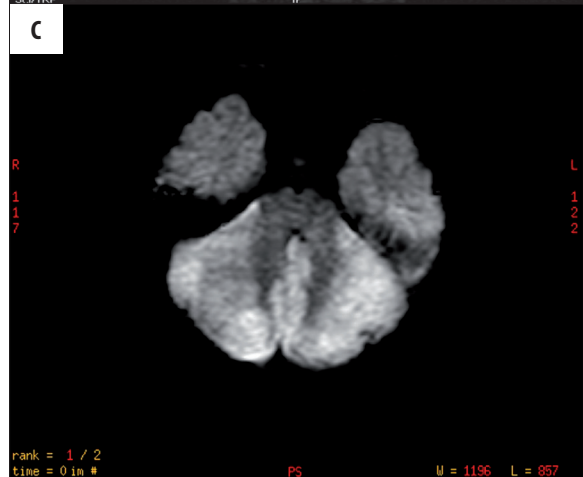
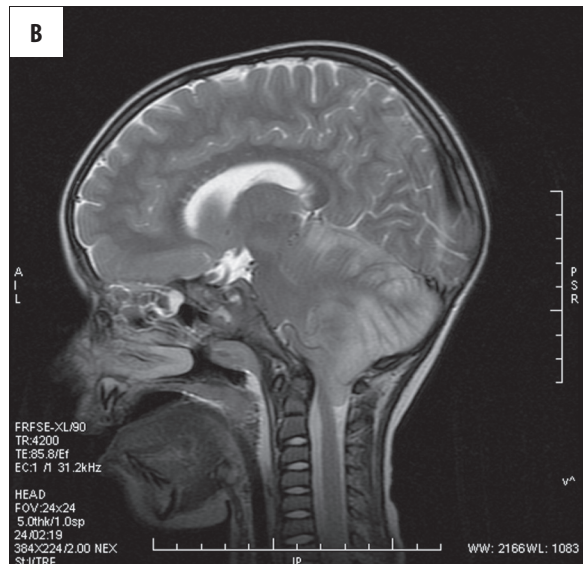
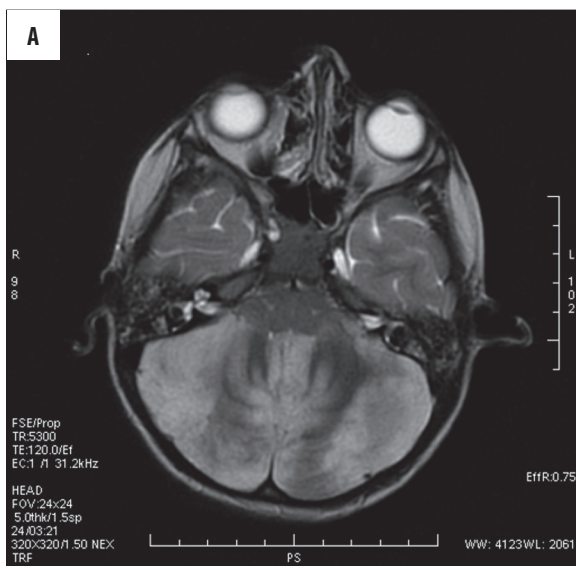


Figure 2. Brain MRI: (A) Axial, (B) Sagittal T2 WI, and (C) diffusion weighted images, showing a massive cortical swelling in the cerebellar hemispheres and the vermis, with increased signal intensity. A significant mass effect with obliteration of the basal cisterns.

After four days of hospitalisation, the boy's clinical condition deteriorated dramatically and he was transferred to the Paediatric Intensive Care Unit (PICU) where he was subjected to an emergent CT of the head (axial scanning, slice thickness of 4 mm) before and after i.v. contrast administration (Figure 1). The examination revealed a decreased density of both cerebellar hemispheres and of the vermis, as well as an increased volume of the cerebellum, especially within its lower hemispheres, with obliteration of the basal cisterns, compression of the pons, mesencephalon and IV ventricle. At the same time, a slight distension of the supratentorial ventricular system was shown. On the same day, due to the severe condition of the child, an emergency MRI was carried out, under general anaesthesia. The brain MRI was performed with a 1.5T scanner in T1-weighted images, before and after i.v. administration of a paramagnetic contrast medium, in T2-weighted images,

FLAIR, DWI-EPI sequences. The examination showed features of a massive, irregular swelling of the cortex of the cerebellar hemispheres and vermis, with a diffuse hyperintense signal in T2-weighted images, in FLAIR and DWI-EPI sequences (Figure 2). After contrast administration, there were no features of enhancement within the changed cortex. Due to the mass effect, the IV ventricle, pons and medulla oblongata were compressed. Mesencephalon and cerebral aqueduct were compressed as well. Cerebellar tonsils were herniated downwards, to the foramen magnum, reaching 16 mm beneath the level of the foramen magnum, obliterating the subarachnoid space and compressing the spinal cord. An upper part of the cerebellar vermis herniated upwards, to the tentorium cerebelli. The supratentorial ventricular system was slightly enlarged; periventricularly there were areas corresponding to a backward absorption of the cerebrospinal fluid to the tissue of the brain.

Owing to the visualised (in the above mentioned examinations) cerebellar oedema with features of herniation,

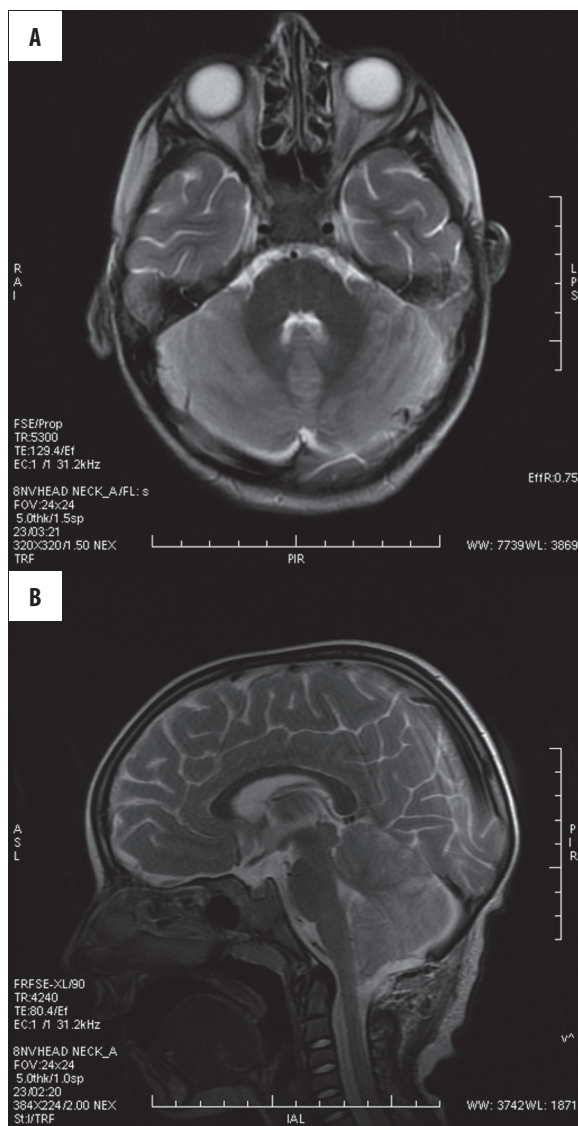


Figure 3. Brain MRI at 6 days after suboccipital craniectomy. **(A)** Axial and **(B)** Sagittal T2 WI showing a regression of the area of increased signal intensity and reduction of signal intensity. A distinct regression of the mass effect.

and due to patient's deterioration, he was subjected to an emergent decompression on the same day (removal of the occipital bone and resection of the C1 vertebral arch). On admission to the Department, the patient's condition was bad but stable. A qualitative and quantitative disturbance of consciousness, hyperaesthesia, aphasia, and paraplegia were found. His circulatory and respiratory system was efficient. After 6 days following the decompressive procedure, a control MRI of the brain was performed (Figure 3). It showed a visible regression of the previously found cerebellar changes – decrease of the diffuse area hyperintense in T2-weighted images, and reduction of signal intensity. The cerebellar swelling regressed substantially: there was no compression on the brain stem or on the IV ventricle, the lower part of the cerebellar vermis moved by more than 1 cm upwards, as compared to the previous examination results, and there appeared cerebellar grooves. The lumen of the supratentorial ventricular system decreased and the grooves of the

cerebellar hemispheres became wider, as compared to the previous test. A slow but significant clinical improvement was observed simultaneously. After a week, the patient regained his full consciousness and remained in a very good emotional contact with his parents; he could hear and understand speech. In the second week of hospitalisation, the patient started talking with single syllables, and then, after a few days, he was uttering simple words and building sentences. Muscle tone in the lower limbs was still decreased; the symptoms of paraplegia were present. Patient's condition was gradually improving and after a 17-day stay at the Infectious Diseases Observatory Department (IDOD), he was transferred to the Neurological Department for further psychomotor rehabilitation. After the rehabilitation, that is three months from the disease onset, the boy had a control MRI before and after intravenous contrast administration (Figure 4). The examination revealed enlargement of the paracerebellar space of up to 1.3 cm (in the width), bilaterally and posteriorly. Similarly enlarged was the fluid cistern around the left cerebellar hemisphere, laterally. There appeared features of a minor, symmetrical cortical atrophy of both cerebellar hemispheres and of the vermis, as well as a slight enlargement of the IV ventricle. No features of oedema or of a pathological contrast enhancement were found. In the DWI-EPI sequence, there were no foci of increased signal intensity, corresponding to a restricted diffusion. One year from the disease onset, the boy is a healthy, mentally and physically fit, child (Figure 5).

Discussion

Cerebellitis is one of the main causes of acute functional disorders of the cerebellum in children. It is characterised by an acute or subacute onset, manifesting itself with cerebellar ataxia during or after an infection or after vaccination. The disease is connected with a wide range of symptoms: from an asymptomatic course to life-threatening complications connected with brain swelling. The most common symptoms include truncal ataxia, dysmetria and headache, as well as nystagmus, dysarthria, wide-based gait, hypotonia, nausea, and vomiting [5,6]. Fever and meningeal symptoms may appear, but not necessarily. The results of the cerebrospinal fluid test may fall within the normal range or reveal pleocytosis [7], as in the case of our patient.

Viral infections of the nervous system are more frequent than the bacterial ones; They are mainly caused by the varicella virus, virus of the measles, mumps or rubella [8]. In less than 1% of cases of acute viral infections, the disease concerns the structures of the central nervous system (CNS). However, it rarely happens that the neuroinfection is located within the cerebellum solely and an isolated massive cerebellar oedema requiring decompression of the posterior cranial fossa is extremely seldom. In most of the cases, the cause of the disease remains unknown [8], as it was also in our patient, because he received a lot of immunoglobulin preparations while being hospitalised at the PICU and thus the serological diagnostics and attempts to establish the aetiology failed. One of the possible causes of the cerebellar swelling was the complication of herpes simplex virus (HSV) infection due to the contact that the boy had one month earlier with his sister who was then infected with recurrent herpes. This is why Aciclovir was introduced.

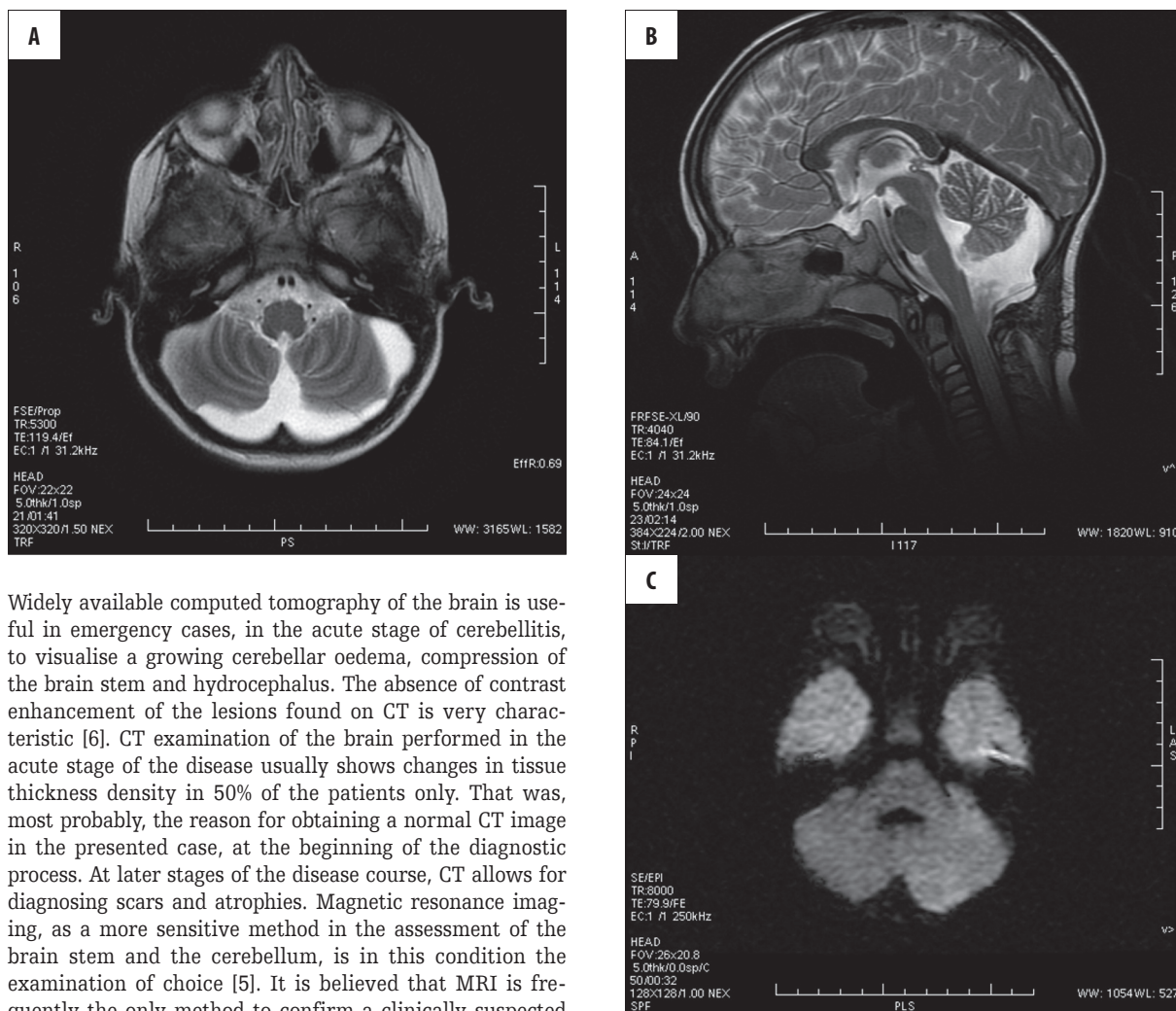


Figure 4. A follow-up brain MRI at three months after the disease onset: (A) Axial and (B) Sagittal T2 WI and (C) diffusion weighted images showing an enlargement of the paracerebellar space and signs of a slight, symmetrical cortical atrophy of both cerebellar hemispheres and of the vermis, as well as disappearance of signs of restricted diffusion.

Widely available computed tomography of the brain is useful in emergency cases, in the acute stage of cerebellitis, to visualise a growing cerebellar oedema, compression of the brain stem and hydrocephalus. The absence of contrast enhancement of the lesions found on CT is very characteristic [6]. CT examination of the brain performed in the acute stage of the disease usually shows changes in tissue thickness density in 50% of the patients only. That was, most probably, the reason for obtaining a normal CT image in the presented case, at the beginning of the diagnostic process. At later stages of the disease course, CT allows for diagnosing scars and atrophies. Magnetic resonance imaging, as a more sensitive method in the assessment of the brain stem and the cerebellum, is in this condition the examination of choice [5]. It is believed that MRI is frequently the only method to confirm a clinically suspected inflammation of the mesencephalon and hindbrain. Lesions within the white and grey matter are hypointense or isointense on MRI in T1-weighted images, while in T2-weighted images and in FLAIR sequence, there is an increase in signal intensity and the lesions may be limited to the cerebellar cortex, with its oedema being the most characteristic MRI image of the disease [6]. Cerebellar involvement can produce different images: most often it is represented by spotted or diffuse lesions in both cerebellar hemispheres, and less frequently by lesions limited to one cerebellar hemisphere only, imitating a tumour and thus postponing the right diagnosis. Normal image is also possible [5,6,9]. There were cases of involvement of the cerebellar vermis and upper peduncles [6,10]. Hydrocephalus resulting from compression of the fourth ventricle and of the basal cisterns by a swollen cerebellum, was present in nearly a half of cases presented in the available literature [4,5].

With termination of the disease course, the cerebellum may return to its original contour or there may be visible features of atrophy. Signal enhancement in T2-weighted images may remain for many months [6]. Even in the absence of hyperintense lesions in T2-weighted images in the course of the disease, a gradual development of cerebellar atrophy may also occur in the late stage of the disease. Features that may facilitate diagnostics of cerebellitis in the late stage include:

distended gyri of the cerebellum, enlargement of the IV ventricle and a slight increase in signal intensity in FLAIR images, as a result of glial proliferation after past disease [4].

In the MRI examination, contrast medium administration results in the enhancement of the cerebellar structures and of the pia mater. However, this is not a regular symptom; in the case presented by us, there was no contrast enhancement. The results of the MRI determine the diagnosis but their prognostic value is limited [5].

In the case presented in this work, the obtained DWI images performed in the acute stage of the disease revealed a diffuse area of increased signal intensity in the cortex of the hemispheres and in the cerebellar vermis. The region was hypotense on the ADC map (apparent diffusion coefficient) and hyperintense in EADC images (exponential apparent diffusion coefficient), which confirmed definitively that there is a restricted diffusion, i.e. cytotoxic oedema



Figure 5. The patient after the disease was controlled, showing no neurological deficits.

caused by acute inflammatory lesions. The DWI method has an advantage over a conventional brain MRI, as it may be the first to show the lesions and be the first symptom of cerebellitis. An increased DWI signal normally resolves with clinical improvement of the patient's clinical condition [12,13]. This was also the case in our patient.

In most of the cases, a symptomatic, anti-inflammatory and antiviral treatment is introduced. Only in unusual situations, it is necessary to carry out an external ventricular drainage or, even more rarely, a surgical decompression of the posterior cranial fossa in case of cerebellitis which may lead to herniation [8,9] (it turned out to be necessary in our case).

Cerebellitis should be differentiated with poisoning (with e.g. cyanide), acute disseminated encephalomyelitis

(ADEM), Lhermitte-Duclos Disease (LDD), extensive infiltrating glioma or lymphoma, vasculitis and demyelinating processes [2,6]. Enhancement of lesions after contrast administration in the MRI examination, as well as regression of the clinical symptoms, allow for exclusion of LDD [5]. It is not always possible to differentiate between cerebellitis and a rare form of ADEM limited to the cerebellum. This is because these two conditions have a similar clinical and MRI picture and are both preceded by infection or vaccination [2,11]. Tumours of the posterior cranial fossa tend to be unilateral and well-delineated from the surrounding tissues; contrast enhancement in the MRI examination concerns tumours within the brain and not the pia mater, and follow-up examinations show no regression of the lesions. Differentiation with demyelinating processes is easier because they tend to occur in the white matter [6].

Cerebellar atrophy requires also differentiation with other causes than the past cerebellitis. They include: hypothyroidism, paraneoplastic syndrome, cerebellar stroke, alcohol or phenytoin overuse and genetic syndromes (SCA – spinocerebellar ataxia, DRPLA – dentarubropallidolulsian Atrophy) [4,14].

Conclusions

Cerebellitis is a rare complication of viral infections and in some seldom cases may lead to swelling of the structures of the posterior cranial fossa and herniation requiring a decompressing suboccipital craniectomy. Due to that fact, a rapid introduction of CT and MRI diagnostic procedures is required to establish the severity of the disease and to apply adequate treatment.

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